Kawasaki Disease
(Also known as Mucocutaneous Lymph Node Syndrome)

May 2003

1) THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent
The cause of Kawasaki disease is unknown but presumably is an infectious or toxic agent.

B. Clinical Description and Laboratory Diagnosis
Kawasaki disease is a multi-system disease primarily affecting children under 5 years old. It is an acute, self-limited, systemic vasculitis characterized by an initial high spiking fever that can persist for 1 to 2 weeks. It does not respond to antibiotics, and it is associated with extreme irritability and mood changes. The fever is accompanied by bilateral conjunctivitis (seen in about 85% of cases); changes to the lips and mouth including dry, red or cracked lips, a sore red throat, and/or strawberry tongue (90%); changes to the extremities including peeling of skin, rashes and/or swelling of the hands and feet (75%); a generalized red rash affecting the trunk or perineal region (80%); and an enlarged cervical lymph node (usually solitary, often unilateral) (70%). Other common symptoms include severe abdominal cramping, diarrhea and vomiting. The convalescent (recovery) phase can be lengthy. Cardiac involvement is a major cause of morbidity and mortality associated with Kawasaki disease. Approximately 20% of untreated cases develop coronary artery aneurysms within 6 weeks. Prompt treatment can reduce aneurysm formation to about 8% of cases, as well as reducing fever and inflammatory signs. The case-fatality ratio for Kawasaki disease is 0.1% to 1%. Other complications can involve any organ of the body.

There are no specific laboratory tests to confirm or exclude Kawasaki disease. Diagnostic decisions are made based on clinical presentations when there are no other reasonable explanations for the illness. Leukocytosis, thrombocytosis and elevated sedimentation rate (ESR) are common findings in the early stage of illness. The ECG changes and coronary artery aneurysms can be observed in the fifth or sixth week of illness.

C. Reservoirs
Reservoirs are unknown.

D. Modes of Transmission
Modes of transmission are unknown.

E. Incubation Period
The incubation period is unknown.

F. Period of Communicability or Infectious Period
The disease is not known to be communicable from person-to-person.

G. Epidemiology
Kawasaki disease occurs worldwide with most cases reported from Japan, where nationwide epidemics have been documented. The estimated number of new cases each year in the United States is approximately 2000. The peak age of occurrence in the United States is between 18 and 24 months, with 80% of cases reported in children younger than 5. Males are affected more frequently than females, and children of Asian descent have the highest incidence rate. Epidemics occur on a 2 to 3 year cycle, usually during the winter and spring. While the mode of transmission for Kawasaki disease has not been documented, siblings (especially twins) of cases are more likely to be affected than the general population. An average of 23 cases of Kawasaki disease is reported annually to the New Jersey Department of Health and Senior Services (NJDHSS).

2) REPORTING CRITERIA AND LABORATORY TESTING SERVICES

A. New Jersey Department of Health and Senior Services (NJDHSS) Case Definition

CASE CLASSIFICATION

A. **CONFIRMED**
A febrile illness of greater than or equal to five days duration, with at least **FOUR** of the following physical findings and reasonable exclusion of other diseases (toxic shock, scalded skin syndrome, scarlet fever, rickettsial diseases, or drug reactions):

- Bilateral conjuctival injection
- Oral changes (erythema of lips or oropharynx, strawberry tongue, or fissuring of the lips)
- Peripheral extremity changes (edema, erythema of palms and soles, or generalized or periungual desquamation)
- Cervical lymphadenopathy
- Rash

B. **PROBABLE**
Not used.

C. **POSSIBLE**
Initially reported on the basis of clinical diagnosis until confirmation is obtained; no possible case classifications are retained.

B. Laboratory Testing Services Available

The laboratory features of Kawasaki disease are nonspecific and nondiagnostic; therefore no testing services are offered at the Public Health and Environmental Laboratories (PHEL).

3) DISEASE REPORTING AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

To identify disease clusters and demographic characteristics.

B. Laboratory and Healthcare Provider Reporting Requirements

The New Jersey Administrative Code (N.J.A.C. 8:57-1.8) stipulates that healthcare providers report (by telephone, confidential fax, over the Internet using the Communicable Disease Reporting System (CDRS) or in writing) of any case of Kawasaki disease to the local health officer having jurisdiction over the locality in which the patient lives, or, if unknown, to the health officer in whose jurisdiction the health care provider requesting the laboratory examination is located.

C. Local Department of Health Reporting and Follow-Up Responsibilities.

1. **Reporting Requirements**
The New Jersey Administrative Code (N.J.A.C. 8:57-1.8) stipulates that each local health officer must report the occurrence of any case of Kawasaki disease, as defined by the reporting criteria in Section 2 A above.

Current requirements are that cases be reported to the NJDHSS Infectious and Zoonotic Diseases Program using the Kawasaki Syndrome Report. A report may also be filed electronically over the Internet using the confidential and secure Communicable Disease Reporting System (CDRS).

2. **Case Investigation**

It is the local health officer's responsibility to investigate a case and complete a Kawasaki Syndrome Reporting form. **When reporting electronically enter collected clinical information into "Comments" section.** Much of the information required on the forms can be obtained from the patient's healthcare provider or the medical record.

a. Use the following guidelines for assistance in completing the form:
   1) Accurately record the demographic information, date of symptom onset, whether hospitalized (and associated dates), outcome of disease, and whether the patient has recurrent Kawasaki syndrome.
   2) Collect the information requested in the "Clinical" section. Ask about duration of fever, presence of conjuctival injection, oral changes, peripheral extremity changes, cervical lymphadenopathy, and skin rash. This information is important in defining a case. Ask the healthcare provider to submit a copy of the medical record or enlist his/her aid in completing these sections of the Kawasaki Syndrome Reporting form.
   3) Ask if toxic shock syndrome, scalded skin syndrome, scarlet fever, rickettsial diseases and drug reaction were excluded.
   4) If there have been several attempts to obtain patient information (*e.g.,* the patient or healthcare provider does not return calls or respond to a letter, or the patient refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as possible. Please note on the form the reason why it could not be filled out completely.

NOTE: If CDRS is used to report, enter collected clinical information into the "Comments" section.

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After completing the case report form, it should be mailed (in an envelope marked "Confidential") to the NJDHSS Infectious and Zoonotic Diseases Program, or the report can be filed electronically over the Internet using the confidential and secure Communicable Disease Reporting System (CDRS). The mailing address is:

NJDHSS
Division of Epidemiology, Environmental and Occupational Health
Infectious and Zoonotic Diseases Program
PO Box 369
Trenton, NJ 08625-0369

4) CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (N.J.A.C. 8:57-1.10)

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

Reported Incidence Is Higher than Usual/Outbreak Suspected

If multiple cases of Kawasaki syndrome occur in a city/town, or if an outbreak is suspected, investigate clustered cases. Identify common factors, such age, school, workplace or activities to help elucidate risk factors. Consult with the Infectious and NJDHSS Zoonotic Diseases Program, at 609.588.7500 to help determine a course of action and, if necessary, to perform surveillance for cases that may cross several jurisdictions and therefore be difficult to identify at a local level.

D. Preventive Measures

None.

ADDITIONAL INFORMATION

A *Kawasaki Syndrome Fact Sheet* can be obtained at the NJDHSS website at <www.state.nj.us/health>.

There is no formal CDC surveillance case definition for Kawasaki syndrome. CDC case definitions are used by state health departments and the CDC to maintain uniform standards for national reporting. For reporting a case to NJDHSS, always refer to the criteria in Section 2 A.

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